

## NOTE ON A CASE OF CHRONIC PROGRESSIVE CHOREA.<sup>1</sup>

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IN Osler's monograph on chorea and choreiform affections, 1894, in the article on Huntington's chorea, some account of the Neiter family is given. So far as was known to the author, only four members of the family had at that time been affected, namely, the mother and three children. One of the sons, Peter, then aged fifty-nine, was at one time a patient in the Johns Hopkins Hospital. This case has been fully reported.<sup>2</sup>

In regard to another son, Nicholas, Osler's article contains the following note: "Nicholas Neiter, aged about forty, blacksmith, living at Edgewood, Hartford County, Md. He was seen for me by Dr. Chas. Simon, who reports that he is evidently subject to the disease, as he displays grotesque inco-ordinate movements of the legs, arms and face. Mentally, too, he is inclined to be childish, and is very emotional. He regards himself, however, as in a condition of perfect health and not affected in any way as his brother Peter."

This patient was admitted to the German Hospital during my service, on the 25th of May, 1895. It was very difficult to obtain information from the patient, who was deaf and whose mental condition was very much impaired. Dr. R. Oppermann, who accompanied him to the hospital, stated that his mental condition varied greatly.

His age was given as fifty four; married; occupation, blacksmith. He had not, however, been able to work continuously for some years, and of late had been unable to do any work at all. Twenty years ago, he received some injury to the back as the result of a fall. From that time, though able to go about and work, he constantly complained of pain in the lower part of the

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<sup>1</sup> Read at the meeting of the Philadelphia Neurological Society, November 25, 1895.

<sup>2</sup> Johns Hopkins Bulletin, Vol. 1.

back. It was impossible to get any accurate information in regard to this injury, to which, however, the patient had constantly referred his troubles. They began insidiously about that time, the arms being first affected. The choreic movements were for a long time slight. Of late years they have been progressively more marked. They are increased by emotion and fright. Never lost consciousness; has not suffered greatly from headache; sleeps well.

Condition on admission.—Patient was a large framed man with fairly good muscles; very little fat. Numerous old scars and a few recent abrasions over the arms and shoulders, the result of injuries received in falling. His expression was blank and stupid; he was unable to converse continuously and replied to questions indistinctly. Memory greatly impaired. He from time to time uttered coarse grunting sounds; and in eating made curious noises with the teeth and lips. There was a little drooling of saliva and almost continuous irregular contractions of the facial muscles with occasional grinding of the teeth. The tongue was protruded with a jerk and quickly withdrawn. It was not tremulous. It was covered with a whitish fur. Desire for food was good. The patient, however, was unable to feed himself. Liquid nourishment only could be administered, all attempts at mastication causing violent irregular movements of the lower jaw and face. Bowels moved daily. There was some loss of control of the sphincters; cardiac impulse felt in fifth intercostal space just within the mammillary line. A faint mitral systolic murmur; lungs normal; area of liver dullness normal; spleen not enlarged; urine pale straw, clear, specific gravity 1018, acid reaction; a trace of albumin; no casts; no sugar. The deafness was marked, rendering it difficult to communicate with him. The pupils were of medium size, reacting to light and on accommodation. There was no nystagmus. The temperature ranged from 99° F. A.M. to 100° or 101° F. P.M. The patient was unable to walk or sit in a chair. On one occasion, during his stay in the hospital, he slipped out of bed at night and crawled on his hands and knees some distance along the ward. His habitual position in bed was upon his back with his arms flexed across his breast, his thumbs flexed, adducted and partly enclosed by the flexed fingers. In this position while the patient was awake there were brief periods of rest, but during the greater part of the time there were irregular slow

movements, consisting of alternate extension and flexion of the fingers, hands and arms, as well as of the legs and feet, the movements being more marked and more frequent in the upper extremities. At the same time there were movements of extension and flexion of the head upon the trunk together with irregular slow rotary movements. Attempts at voluntary motion greatly increased the irregular movements. They also became intensified at the time of defecation or the passing of water. During sleep these movements ceased, with the exception of slight twitchings of the muscles of the lower part of the face. The movements were for brief periods under the control of the will. During the examination he could arrest them for periods of fifteen or twenty seconds, the return of the movements being first manifest in the lower extremities. On two occasions violent general convulsions occurred. The knee-jerks were markedly exaggerated. Ankle clonus was not obtained. There was difficulty in determining other reflexes. Sensation was not impaired.

During the night of June 2, the eighth day of his stay in the hospital, the weather being intensely hot, his temperature suddenly rose to 104° F. Great prostration developed, with some abdominal pain. It happened that one of his daughters visited the hospital the next morning and insisted upon at once removing her father to his home in Maryland. The danger of such a course was pointed out to her by the chief resident physician, Dr. Frese. Notwithstanding his remonstrance the patient was placed in the ambulance to be taken to the station. His death occurred in the ambulance a few minutes after leaving the hospital.

Efforts to obtain a post-mortem examination of the body were without success.

I have learned from Dr. Oppermann that Nicholas Neiter had five children. Of these the oldest, a son, aged thirty-four has some irregular motor symptoms, particularly noticeable in his manner of carrying his head, and in jerking about the eyelids. This man is in other respects in good health. The youngest child, also a son, now twenty-two years old, has well-marked irregular muscular movements of the upper extremities. These movements began about a year ago. This young man is stated to have "a great deal of temper, which gives much trouble," otherwise he is hearty and well and has a good digestion. He does not appear to be annoyed by the

muscular movements. The three daughters do not show any evidences of chorea. They suffer occasionally from neuralgic attacks, and one of them has a dry, hacking cough. It is stated that "some of Mr. Nicholas Neiter's kin died of consumption at the ages of 22, 24 and 27.

The number of cases of chronic progressive chorea reported in the literature of medicine is now considerable. Several cases have been reported to this Society. I should have thought it unnecessary to read the foregoing note had it not been that the patient was a member of a family in which the disease is now known to have shown itself in the third generation.